Bullous Cases

Case 1

A 25 years old male presented with extremely itching excoriated papules & blisters which arise on normal or reddened skin. These tend to be distributed symmetrically & in groups. The patient is suffering from abdominal discomfort.

a) What is the most likely diagnosis?

b) Describe the histopathological features?

c) What is the immunofluorescence pattern?

Answer:

a) Dermatitis herpitiformis (DH)

Key points:

- Extremely itching
- Symmetrically & in groups
- Abdominal discomfort

b) Subepidermal vesicle with papillary neutrophilic microabscesses within dermal papillae.

There is ermal perivascular neutrophilic infiltrate, many of which show disintegration of their nuclei (nuclear dust). By EM , skin cleavage occurs within lamina lucida.

c) DIF: granular IgA deposits at dermal papillae, IIF: negative

Case 2

A 23 years old woman presented with a three years history of recurrent blistering eruption that is associated with mild itching. Physical examination: there were multiple, tiny, vesicles on erythematous base, occurring in crops, on trunk & limbs. The distribution was not limited to the extensor aspects. Skin biopsy: there were patchy focal accumulations of neutrophils & scanty eosinophils with fibrin at dermal papillary tips.

a) What is your diagnosis?

b) What is the differential diagnosis?

c) Describe the main pathogenic events?

d) Describe the management plan?

Answer:

a) Dermatitis herpitiformis (DH)

Key points:

- Itching
- Multiple, tiny, vesicles
- Occurring in crops
- Neutrophils at dermal papillary tips.

b) other bullous diseases

c) Defective mucosal berrier allows the passage of macromolecules ofas gluten to the lamina propria evoking an immune response with the production of IgA ab & immune complexes.

Circulating IgA immune complexes are deposited in the dermal papillae & release of chemoattractants. Neutrophils are recruited to upper dermis &release proteases leading to intra-lamina lucida separation & vesicle formation.

d) dapson 100-200 mg /day, gluten free diet

Case 3

A 60 years old male presented with multiple small tense bullae on erythematous skin of 2 years duration. Some of the bullae were preceded with urticarial wheals with some itching. The oral cavity was free except from geographic toung.

a) What is the most likely diagnosis?

b) Describe the histopathologic picture?

c) What is the immunofluorescence pattern?

Answer:

a) bullous pemphigoid

Key points:

- 60 years old (old age)
- Tense bullae
- Urticarial wheals with itching
- Oral cavity free

b) Subepidermal bullae with predominant eosinophils

c) DIF: linear IgG &/or C3 along DEJ , IIF: IgG ab against BP antigen at DEJ

Case 4 (Diploma 11/2011)

A 35 years old male suffering from painful oral ulcerations for the last 3 months, he developed flaccid bullae containing clear fluid over the trunk & proximal parts of his extremities one week ago. The bullae developed on both normal & erythematous skin. They rupture easily with painful erosions.

a) What is the most likely diagnosis?

B) How can you confirm your diagnosis?

c) How can you manage this case?

Answer:

a) Pemphigus vulgaris

Key points:

- 35 years old (middle age)
- Painful oral ulcerations
- Flaccid bullae
- Rupture easily

b) Biopsy: histopathlogy: suprabasal acantholytic bullae, tombstone appearance, intraepidermal eosinophilic microabscesses, acantholysis involves hair follicles.

DIF: Intercellular IgG &/or C3, IIF: IgG AB against desmosomes

c) see the guidelines

Case 5 (Master 4/2005)

A male patient 40 years old presented with scaly, crusted cutaneous erosions, often on an erythematous base. They are usually well demarcated & have a seborrheic distribution. He does not have clinically apparent mucosal involvement even with widespread disease. He complains of burning & pain in association with the skin lesions. Histopathological picture reveals clefting in the upper spinous layer of the epidermis with several acantholytic cells detaching from the roof of the blister. In the active state of this disease direct immunofluorescence demonstrates 100% IgG deposition on the keratinocyte cell surfaces.

1- What is the diagnosis of this case?

2- Comment on : direct & indirect immunofluorescence in pemphigus vulgaris & pemphigus foliaceus?

3- Write about the differences between direct & indirect immunofluorescence?

4- Mention another specific method used to demonstrate the target molecule of the previous case?

5- Prescribe the treatment of this case?

Answer:

1- Pemphigus foliaceus

Key points:

- Seborrheic distribution
- No apparent oral lesion (exclude PV with painful oral lesion)
- Clefting in the upper spinous layer
- IgG deposition on the keratinocyte cell surfaces.

2- PV: DIF: Intercellular IgG &/or C3, IIF: IgG AB against desmosomes

PF: DIF: Intercellular IgG &/or C3, IIF: IgG AB against desmosomes

3 DIF: detects in vivo antibodies bound to tissue antigens in perilesional skin. IIF: detects circulating serum antibodies (substrate sections react with serially diluted serum from patient incubated with anti-IgG or other specific fluorescent dye-tagged antibody). Best substrate: Monkey esophagus (PV), guinea pig esophagus (PF)

5- ttt: topical steroid, prednisone 1mg/kg with or without steroid sparing agent (dapsone, gold, hydroxyl-chloroquine , azathioprine)

Case 6 (Master 4/2005)

Female patient 45 years old developed pustules mainly in axillae, groins & on flexor aspects of limbs. The pustules showed annular arrangement. The pustules were flaccid with pus accumulation in the lower half of the pustule. No oral lesions. The pustules were sterile on culture. The patient had IgA paraproteinemia. Biopsy revealed subcorneal pustules containing neutrophils with occasional eosinophils. A few acantholytic cells were found in the base of the pustules.

The dermal papillae contain dilated capillaries with perivascular infiltrate composed of neutrophils. Direct immunofluorescence is negative.

1- What is your diagnosis?

2- What is the differential diagnosis?

3- What is the treatment of this case?

4- What is the prognosis of this case?

Answer:

1- Subcorneal pustular dermatosis

Key points:

- Female patient 45 years
- Pustules in axillae, groins
- Annular arrangement
- flaccid with pus accumulation in the lower half of the pustule (hypopyon level)
- No oral lesions
- IgA
- Dermal papillae contain neutrophils
- Direct immunofluorescence is negative

2- other bullous diseases

3- ttt: dapsone 50-150 mg/day

4- Chronic relapsing disorder, healing occurs with superficial crust & later on with brown pigmentation.